Case Report

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A Case of Acute Coronary Syndrome Under Immunosuppression Who is the Criminal Neutrophils or T Cells?

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Abstract

Chronic granulomatous disease (CGD) is a primary immunodeficiency characterized by recurrent, life-threatening bacterial and fungal infections of the skin, airways, lymph nodes, liver, brain, and bones. These infections most commonly occur in organs in contact with the outside world (lungs, gastrointestinal tract, and skin), as well as in lymph nodes that drain these structures. While involvement can be seen in many organs, there is no known cardiovascular involvement. Our case is an ACS case that has a different place in the literature because acute coronary syndrome (ACS) was seen in a twenty years old male patient with a diagnosis of chronic granulomatous disease.

Keywords: Acute coronary syndrome, bone marrow replacement therapy, chronic granulomatous disease, coronary angiography, graftversus host disease

Introduction

Chronic granulomatous disease (CGD) is an immunodeficiency syndrome characterized by recurrent, life-threatening bacterial and fungal infections of the skin, airways, lymph nodes, liver, brain, and bones. These infections most commonly occur in organs in contact with the outside world (lungs, gastrointestinal tract, and skin), as well as in lymph nodes that drain these structures. These children are normal at birth. However, infections can be quite severe in the early period. Chronic granulomatous disease is genetically inherited. Although initially thought to have only an X-linked form of inheritance, its discovery in girls in 1968 enabled the recognition of autosomal recessive forms. (1) Although incidence rates are higher in geographies where consanguineous marriage is more common, CGD is thought to affect one in 20000 to 250000 live births without ethnic preference. (2-5) As an immunodeficiency syndrome, it causes severe persistent bacterial and fungal infections such as tuberculosis, aspergilloma and osteomyelitis. (6-7) In disease management, it is aimed to quickly identify and treat acute infections and to prevent secondary granulomatous complications, together with prophylactic antibacterial, antifungal and immunomodulatory drugs. Although hematopoietic stem cell transplantation appears to be the only widely available curative treatment for patients with CGD, recent advances in gene therapy may also provide a safer and more direct option. (8) However, bone marrow transplant cannot be a definitive solution and graft versus host disease (GVHD) can be seen in patients. Although the risk of transplant related mortality and GVHD is high, survival in children has been shown to be significantly higher. The probability of developing GVHD with the increase in age has been found to be higher in studies. (9)

Case Report

A 20 years old male patient with a known diagnosis of chronic granulomatous disease, who underwent bone marrow transplant in 2008 and 2019 and was followed up in pediatric immunology with the suspicion of graft versus host disease, was consulted from the emergency department with the complaint of chest pain in the retrosternal region. From the anamnesis, it was learned that the patient was followed up with the diagnosis of chronic granulomatous disease, the last bone marrow transplant was performed in 2019, and immunosuppression treatment was started a week ago with

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the diagnosis of graft versus host. When his family history was questioned, it was learned that his sister also died at a young age due to chronic granulomatous disease. The patient applied to the emergency department with the complaint of compressive chest pain in the retrosternal region. In the drugs he uses, methylprednisolone 16 mg tablet, ursodeoxycholic acid 200 mg tablet, calcium carbonate 1000 mg effervescent tablet, cyclosporine 25 mg tablet, mycophenolate mofetil 500 mg tablet, posacanazol 100 mg tablet, zinc sulfate 50 mg tablet, Cetirizine dihydrochloride 10 mg tablet, pantoprazole 40 mg tablet, levofloxacin 500 mg tablet, acyclovir 800 mg tablet were seen. It was learned that he had no history of smoking and dyslipidemia. On physical examination, blood pressure was 110/60 mmHg and saturation was %95 , heart rate was 116/min. Electrocardiography (ECG) of the patient with typical anginal symptoms showed normal sinus rhythm and diffuse ST elevations. (figure 1) Echocardiography was performed on the patient. The ejection fraction was %40 anterior, and wall motion defect in the septum and first degree mitral regurgitation were observed. In laboratory results, troponin is 5.39 ug/l, ck-mb is higher than 300 ug/l, creatinine normal alanine aminotransferase (ALT) and aspartate aminotransferase are high, C-reactive protein (CRP) is within normal limits, hemoglobulin 19.6 g/dl, platelet 190 thousand, white blood cells was seen as 12 thousand. The patient, who had acute coronary syndrome findings on ECG and ECHO, describing typical anginal symptoms, had elevated troponin and was taken to coronary angiography (CAG) with a preliminary diagnosis of ACS. The right coronary artery (RCA) was normal, the proximal circumflex (CX) lesion was %70, and the left anterior descending artery (LAD) proximal was %100 occluded. The total occluded lesion proximal to the LAD was predilated with a 2.5*30 mm balloon, and then a 2.5*33 mm drug-coated stent was implanted. Then, the stent was post dilated with a 2.75*12 mm balloon. Full disclosure has been achieved and the process has been terminated. (Figure 2) The patient was taken to the coronary intensive care unit. Dual antiaggregant therapy was started. The patient with elevated hemoglobin levels was consulted to hematology. One unit of phlebotomy was performed and mycostatin oral suspension was started. The patient, who was followed in isolation in the coronary intensive care unit for two days, was discharged with full recovery after medical treatment was arranged.

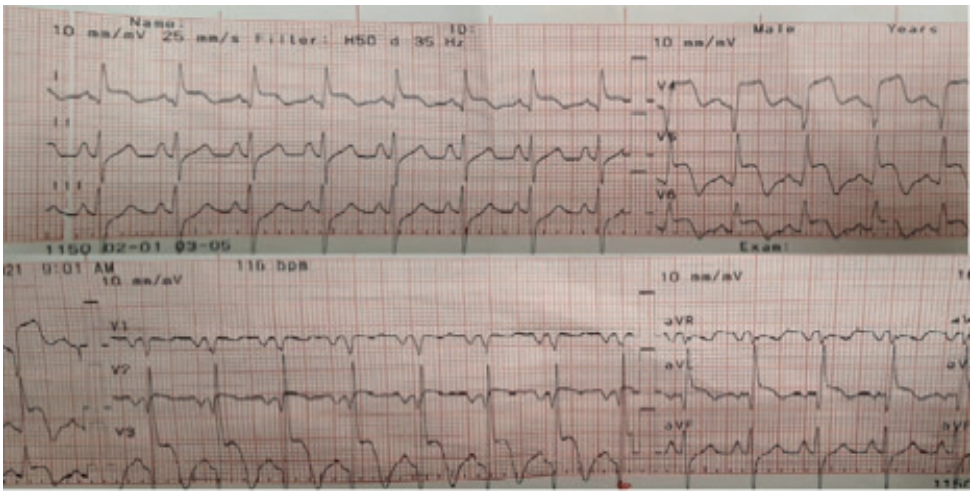

Figure 1.

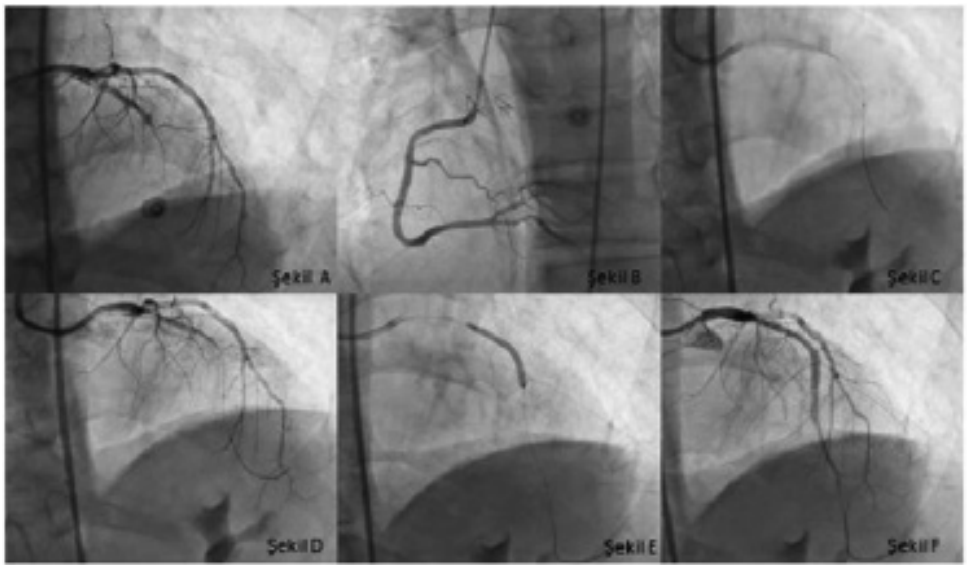

Figure 2.

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Discussion

Chronic granulomatous disease is a disease that affects the immune system and progresses with the formation of granulomas as a result of the inability of microorganisms to be phagocytosed by neutrophils. Many organs involvement can be seen, but cardiovascular involvement is unknown. However, there are also granulomatous diseases with cardiovascular involvement. When we look at the literature, we see examples of this. For example, acute coronary syndrome and left anterior descending artery occlusion similar to our case were detected in a young male patient with Wegener's granulomatosis. (10) In another example, in two different patient groups with Takayasu artery and rheumatoid arthritis, early atherosclerosis was observed in those with Takayasu arteritis.(11) This was thought to be because atherosclerosis was associated with chronic systemic inflammation without cardiovascular risk factors. It has been accepted that vascular remodeling and endothelial activation play a role in the development of atherosclerosis in autoimmune granulomatous diseases. Considering the classification of myocardial infarction, type 1 myocardial infarction develops due to atherosclerotic plaque rupture, while type 2 myocardial infarction develops due to an ischemic cause due to an imbalance of myocardial oxygen supply and demand. In the light of all these, our patient was evaluated as type 1 myocardial infarction in the presence of ST elevation on ECG, elevated cardiac biomarkers and anginal symptoms. Although antibiotics, intravenous immunoglobulin and bone marrow transplantation are used in the treatment of chronic granulomatous diseases, GVHD can be seen. In cases where GVHD develops, the only treatment that can be done is to suppress the immune system. For immunosuppressive treatment, drugs such as methylprednisolone, cyclosporine, mycophenolate mofetil are usually used in the first stage. Although macrophages are in the first place in the pathogenesis of atherosclerosis, both CD8 and CD4 T lymphocytes are seen at every stage of atherogenesis. The presence of these cells in the atherosclerotic lesion suggests that atherosclerosis occurs as a result of an immune or perhaps an autoimmune response. As a matter of fact, in a study conducted in the literature, subendocardial ischemia was observed in the imaging performed with adenosine in patients with Wegener's granulomatosis, and it was shown that myocardial ischemia disappeared after cyclophosphamide and steroid.(12) Considering all these studies and our case, the increased number of neutrophils in granulomatous diseases and the high number of T cells brought about by an autoimmune mechanism raises the question of whether neutrophils should be responsible for the etiology of atherosclerosis or whether T lymphocytes should be responsible. Our case enters the literature as a special case emphasizing this contradiction.

Informed consent: *Informed consent was obtained from the patient for the publication of the case report and the accompanying images.*

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